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Contemporary Perspectives on Cochlear Implants in Children, presented in partnership with Cincinnati Children's

Presenter: Jill Huizenga, AuD, CCC-A;
Michael Scott, AuD, CCC-A, FAAA

Moderator: Carolyn Smaka, AuD, Editor in Chief, AudiologyOnline

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Contemporary Perspectives on Cochlear Implants for Children

Jill Huizenga, Au.D., CCC-A
Michael Scott, Au.D., CCC-A

Objectives

- Describe current pediatric cochlear implant candidacy and the evaluation process.
- Provide case studies to highlight situations where current candidacy guidelines are not always met.
- Identify important aspects of an evaluation protocol and management of pediatric patients who receive cochlear implants.

Disclosure:

Permission to share case studies obtained from patients and parents, but have been highly modified

Cochlear Implant Team at CCHMC

- Audiology
- ENT
- Speech Pathology
- Social Work
- Developmental Pediatrics

The Team Approach:

- Evaluation and input from several disciplines
- A collaborative decision made on every case
- ChIP (modified from Hellman et al, 1991):
  - Objective **tool** for evaluating potential cochlear implant candidates
  - Criteria to determine areas of "no concern," "mild to moderate concern," and "great concern"
  - The team meets to discuss their finding for each child evaluated
  - The team members complete the Children's Implant Profile (ChIP)
  - Recommendations to proceed with surgery or for other services are made following the completion of the ChIP

Factors that Affect Outcomes

- Previous auditory experience
- Age at implantation
- Duration of deafness
- Presence of other disabilities
- Anatomy
- Presence of language
- Having sufficient motivation
- Good support system
- Consistent hearing aid use

FDA Criteria

- Adults (18 years and older)
  - Ranging from moderate to profound sensorineural hearing loss bilaterally
  - Limited benefit from appropriately fit binaural hearing aids
    - Ranging from <40% HINT to ≤60% on any speech perception test in the best aided condition)
- Children (12 months to 17 years, 11 months)
  - Profound bilateral loss (ranging to severe to profound for 2+years)
  - Limited benefit from appropriate fit binaural hearing aids
    - Younger children: Lack of progress on auditory milestones with appropriate amplification and enrollment in intensive auditory rehabilitation
    - Older children: Ranging from ≤ 20% MLNT/LNT to <12% PBK and <30% HINT

Special Populations

- Studies have shown that 40-50% of children with hearing loss will have an additional disability. (Wiley et al, 2004)
- It is important that realistic and appropriate expectations are discussed and understood by the family and professionals involved.
- Since it is ideal to implant a child at an early age, there will be children who receive implants prior to the identification of additional disability.
- Disabilities such as autism or apraxia may not be identified until a child is 2-4 years of age.
- A language, learning, or cognitive disorder will still be present after a child gets a CI. It is important that everyone understands that the implant is not going to resolve all issues.

Special Populations:
Evaluation for Cochlear Implants

- Challenges:
  - Obtaining accurate audiometric information
  - Understanding family expectations
  - Available resources
- Tools:
  - Objective measures (ABR, ASSR, OAE, etc.)
  - Speech perception- not always possible to obtain, much less with great reliability
  - Questionnaires and Profiles (IT-MAIS, ASC, etc.)

Other Considerations:

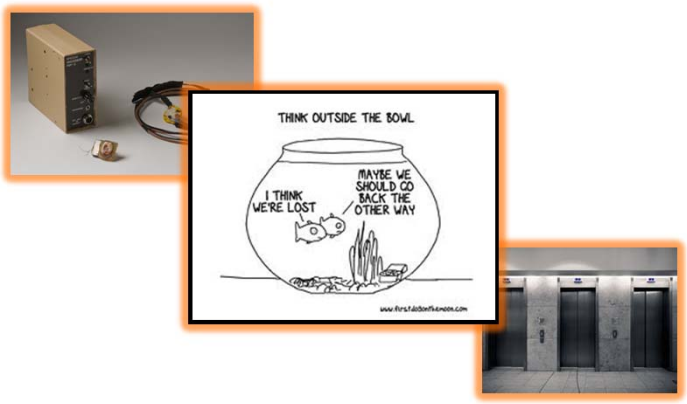
- Evaluate for communication ability, not just hearing sensitivity
- Does this child make use of the information he receives from his intact sensory modalities?
  - Environmental involvement – vision and touch
  - Does the child accept or reject this input?
  - How might this relate to tolerance of device wearing or the stimulation it provides?

Special Populations

- Children with additional disabilities *need* an experienced multi-disciplinary team to assist in determining appropriate expectations

- These children <u>can</u> benefit from the evaluation whether or not they proceed with a CI as they will receive a developmental evaluation and appropriate educational recommendations

Case 1: Thinking Outside of the "Box"



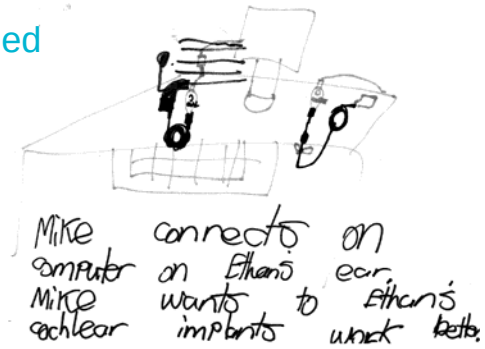
Case 1: Autism and Failed Devices

- 5 year old boy with Autism
- Bilateral profound SNHL secondary to CMV
- Bilateral implantation- was doing well until one day where he suddenly refused to wear either device
- Had to sort out resistance:
  - One implant vs both
  - Which device?
  - Absolute refusal to connect to the computer
  - What to do next?

Programming Tips and Tricks "Environmental Responsiveness"

- **The tool:** go to where the patient is more comfortable
- The child may give you more feedback and/or be more willing to play listening games in a familiar environment such as:
  - the place he has weekly therapy
  - school environment
  - elevator? - in jest, but think outside the box!

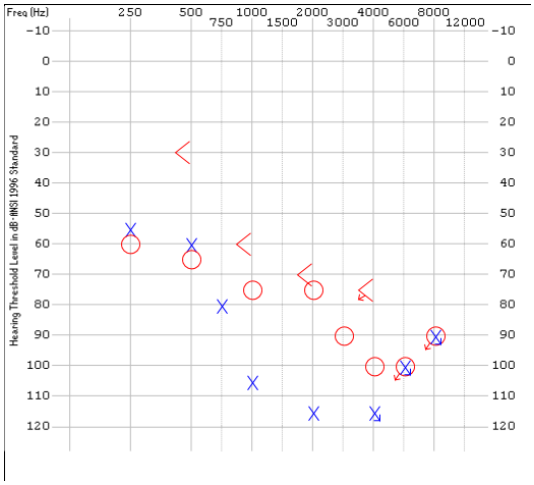
Case 1: Autism and Failed Devices



Case 2: Poor speech and language development

- Pendred syndrome and bilateral EVA with asymmetrical, progressive sensorineural hearing loss
- Not making expected progress with speech and language development despite enrollment in an auditory verbal preschool program
- Questions arose about potential interference from the left (poor hearing) ear

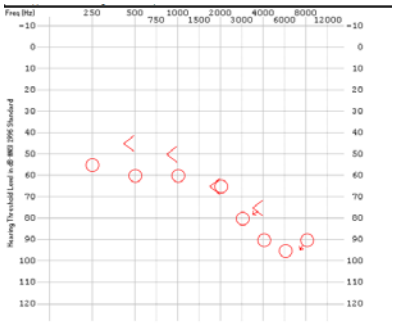
Case 2: limited speech and language development

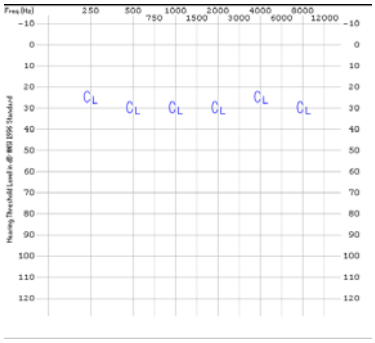


Pre-Op Aided Speech Perception	Right	Left
SAT	20 dB HL	35 dB HLT

Ling 6 Sounds (Right HA)	35 dB HL
/a/ "ah"	Detected
/u/ "oo"	Detected
/i/ "ee"	Detected
/S/ "sh"	Detected
/s/ "ss"	Detected
m/ "mm"	Detected

Case 2: Limited speech and language development **(4 months post-implant)**





Aided Speech Perception	Right	Left
SAT	30 dB HL	20 dB HL
NU-CHIPS	88%	72%

Case 3: visual impairment

- 6 year old girl
- Profound bilateral hearing loss secondary to CHARGE
  - coloboma, heart defect, choanal atresia, retarded growth and development, genital abnormality, and ear abnormality
- Complex inner ear anatomy
  - Incomplete insertion of the electrode array
  - Which portions are in contact with neural structures?
- Severe visual impairment

Case 3: visual impairment

- **Every case is different**
- **Need to scrutinize every channel**
  - One bad channel can mean the difference between a child willing to wear his device vs. constantly taking it off!
- **Need to measure T *and* C levels**
  - Tools for either of these?

Visual impairment: the tools

Visual Reinforcement

- May have to dim the lights significantly
- Light up toys for VRA placed in very close proximity

Conditioned play

- Use toys that entertain, and have lots of them!
- balls and blocks may be less tactile than stars→
- Light up pointer/pen/flashlight/tap-light that the child can turn on when they hear sound

Case 3: visual impairment

- Likes to hear- now teaching him how to replace his own coil with help of hook-n-loop material
- In regular speech therapy: using some signs, but needs max cuing, considering an AAC trial
- continues to turn to his mother when he hears her voice
- responded to different noises being made in his environment and moved his head to look for them in more than 60% of attempts.
- will often localize to someone's voice, but it is not necessarily always when his name is stated

Trends in age at Cochlear Implantation

Pushing the envelope:

- Initially, adults only
- Then 2-18 year old children
- Then 18 month old children
- Now 12 month old children
- Younger with special cases (such as meningitis and risk of cochlear ossification)

Tech: A "culprit" behind the shift

- Advancements in technique and technology
- Universal newborn hearing screening
  - Now, we know almost immediately after the baby is born
  - Baby can be fit with amplification quicker
  - Amplification is verified through objective means
  - Earlier determination of limited benefit
- Amplification and implant technologies are constantly improving

Bottom line questions:

- When does patient performance with a cochlear implant exceed what they currently do with optimized hearing aids?
- CI technology is rapidly improving, but so are hearing aids- length of trial period?
- What are the key audiologic factors?
- How can we be certain that we have all of the (most accurate) information?
- Do the benefits exceed the risks?

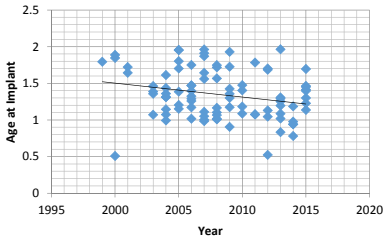
Implantation younger than 12 months?

Cosetti, M., & Roland, J. T. (2010). Cochlear implantation in the very young child: issues unique to the under-1 population. *Trends in amplification*, *14*(1), 46-57.

- Evidence suggests a higher rate of receptive and language development in children implanted under the age of 1
- Outcomes data in auditory perception and linguistic development suggest that early-implanted children may be more likely to achieve their full potential and may reduce or eliminate the need for them to "catch up" or learn at a faster than normal rate to achieve age-appropriate norms

The Trend at CCHMC

- Over 700 children implanted
- Since 2000, those implanted before 12 months: 12
  - Sequential bilateral: 5
  - Simultaneous: 7
  - most within the last few years



Case 4: the ideal candidate

First have to talk about the older sister:

- Identified with profound loss
- Unknown etiology
- Started using sign language at home
- Not implanted until 27 months of age due to insurance issues
- Enrolled at local Oral School
- Immediately "took off" in auditory, speech and language acquisition

Case 4: the ideal candidate

Younger sister:

- Identified hearing loss very shortly after birth
- Genetic testing: Usher syndrome (type 1B)

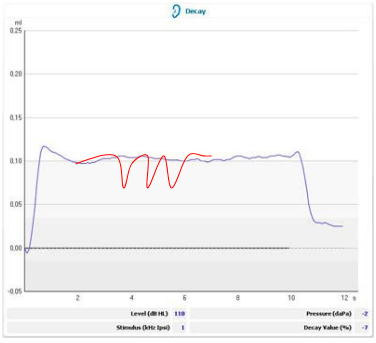
- Vestibular battery: no vestibular function (in physical therapy)
- Quickly fit with appropriate amplification
- Enrolled in early intervention, speech and aural habilitation therapies
- Monitored consistently by multiple providers
- No benefit from traditional amplification as determined by all of her therapists, audiologist, early interventionist, etc.

Cochlear Implants- how to program that young?

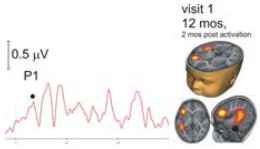
- Implanted at 9 months of age
- Simultaneous bilateral

- First programming:
  - Behavioral observation
  - Objective measures:
    - telemetry
    - eSRT- cumbersome, but worth it

- Follow-up programming:
  - Slightly more frequent initially
  - Feedback from parents
  - Feedback from early intervention



Case 4: the ideal candidate



- Now 4 years old
- Performing on par with, or above age-matched peers
- Word recognition ability at 88 and 96% (PBK), but may be higher due to known vocabulary
- Longitudinal cortical measures to investigate developmental/neuroplastic trajectories that may predict how well a CI user will perform in speech perception tasks
- PLS-5 used to assess auditory comprehension and expressive communication skills at specific time intervals
  - At the chronologic age of 4 years, 1 month:

	Raw Score	Standard Score	Percentile Rank	Age Equivalent
Auditory Comprehension	49	103	58	4 years, 5 months
Expressive Communication	49	104	61	4 years, 6 months
Total Language	98	104	61	4 years, 5 months

*Mean Standard Score = 100,+/- 15

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Case 5: not so ideal?

- Special circumstance
- Meningitis contracted at 4 months of age
- Ossification noted on CT scan
- Implanted bilaterally at 6 months
- Full electrode array insertion, despite ossification noted earlier

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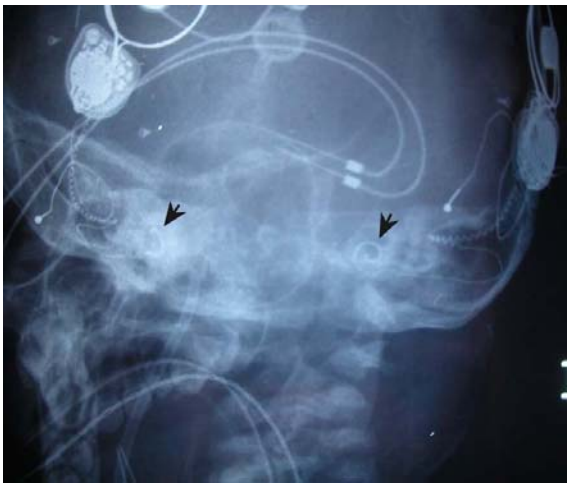
changing the outcome together



Case 5: early implant due to meningitis

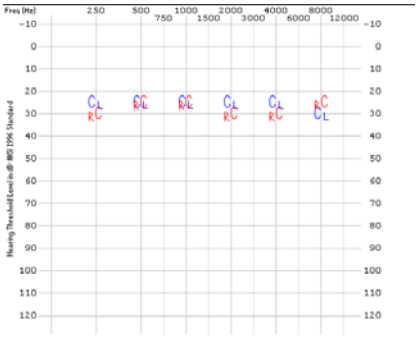
- First programming:
  - No measurable neural telemetry
  - Settings based solely upon behavioral observation
  - Did observe a response to sound initially, but quickly habituated
- Subsequent programming:
  - More frequent follow-up
  - Relied heavily on parent and therapist report

Case 5: early implant due to meningitis

- **Issues**
  - Parents had little to no time to process what hearing loss and implantation truly meant for their child
  - We did not know until later that she would have developmental delays not solely related to hearing loss, but would impact outcomes
- **Today**
  - 7 years old
  - Can complete open-set word discrimination tasks that are age-appropriate
  - Making excellent progress in school, though still has some delays

Case 5: early implant due to meningitis

	Right Cochlear Implant	Left Cochlear Implant	Binaural
Discrimination of Average Speech: % of Words Correct 50 dBHL in Quiet Word List: LNT LNT	92	88	
Discrimination of Average Speech: % of Phonemes Correct 50 dBHL in Quiet Word List: LNT LNT	96	88	
Sentence Recognition in Quiet: % of Words in Sentences Correct 50 dBHL in Quiet Sentence List: HINT-C			51% This is likely an under-estimate of ability due to language-processing issues



Case 6: Case of the Intermittent Implant

- 7 year old boy
- Profound bilateral hearing loss
- Unknown etiology
- Developmentally delayed
- Sequentially bilaterally implanted
- Doing well, until parent calls one day… quite flustered.

"The implant won't stay on!"

Case 6: Case of the Intermittent Implant

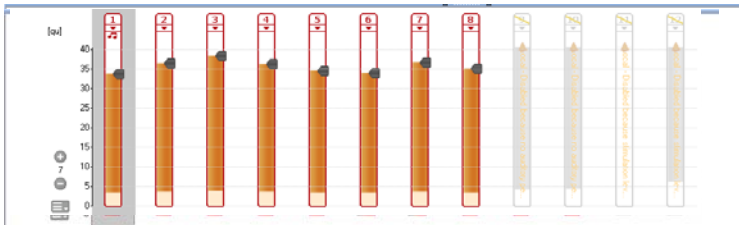
"The implant won't stay on!"

- What does "on" mean?
- What do we suspect first?
- Processor checks out, what next?
- Connected to the computer, and…
- I see the implant connect… sorta…

Case Study 7: Moving target

- 7 year old girl
- Progressive sensorineural hearing loss
- Simultaneous bilateral implantation
- Doing well with her implants, auditory skill development, speech, school, etc.
- New complaints about 2 years after implantation, of not being able to hear /s/ out of the right implant only

Case Study 7: Moving target

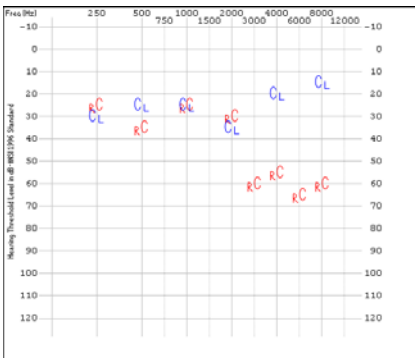


Case Study 7: Moving target

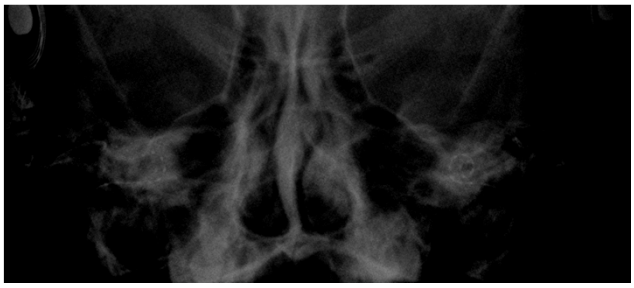
Straw the broke the back: could not maintain good high frequency access

Tool #1: CT Scan

Speech Perception Testing	Aided: Left	Aided: Right
Identification of Speech: Speech Reception Threshold (dBHL)	25	25
Discrimination of Speech: % of Words Correct at 50 dB HL in Quiet (PBK)	80	36



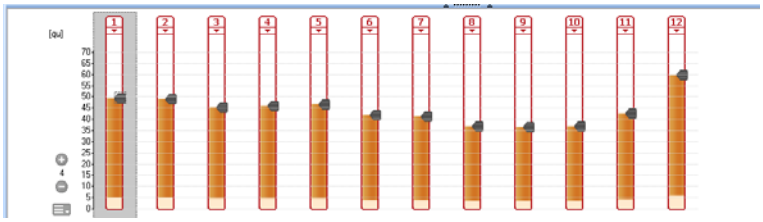
Case Study 7: extruding electrode array



Case Study 7: Surgeon's notes

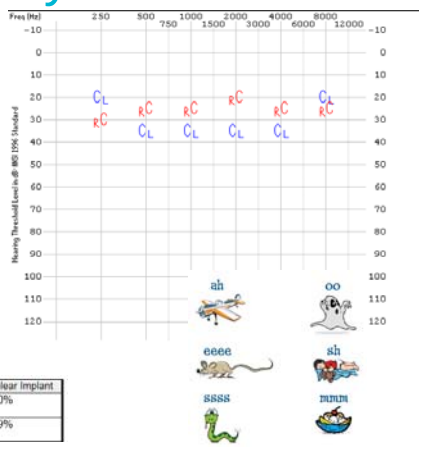
- The electrode was identified as it exited the receiver/stimulator and traced through scar tissue in an anterior direction.
- Please note that freeing the soft tissue from the electrode took approximately 3 hours and was extremely difficult secondary to adhesive scar tissue and required slow and meticulous dissection so as not to damage the electrode array.
- The facial recess was completely filled with scar tissue, and it became evident upon dissection that the electrode array was not present within the cochlea at the time of dissection.
- After removal of the vast majority of the scar tissue from the electrode and after completely cleaning, the portion of the electrode could be re-inserted into the cochlea.

Case Study 7: Moving target no longer moving!



Case Study 7: today

- Stability!
- Programming, thresholds, Lings, etc.
- Still states that the left is his favored ear
- Tool: Ling 6 sounds

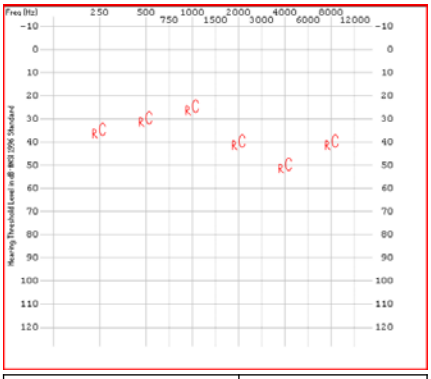


Speech Perception Testing Results	Bilateral Cochlear Implant	Left Cochlear Implant	Right Cochlear Implant
Discrimination of Average Speech: % of Words Correct 50dBHL in Quiet		92%	80%
Discrimination of Average Speech: % of Phonemes Correct		97%	89%

Case 8 Stapes Gusher

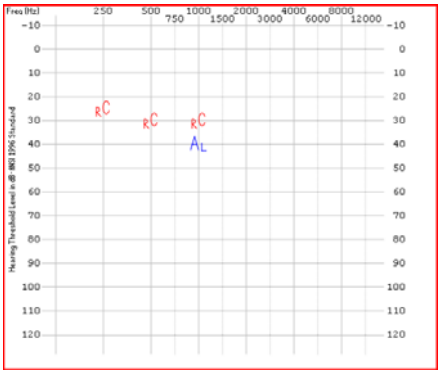
- Etiology: X-Linked Stapes Gusher
- Age at identification: 1 month
- Age fit with hearing aids: 3 months
- MRI Results: There is a symmetric appearance to the internal auditory canals and cochlea bilaterally. **The internal auditory canals have a bulbous configuration with incomplete separation of the fundi of the internal auditory canals with the basal turn of the cochlea. This causes a corkscrew-like appearance to the cochlea and adjacent internal auditory canals. The modiolus is absent and the cochlea is incompletely partitioned bilaterally.** The labyrinthine segment of both facial nerve canals are mildly prominent in relation to the remaining course of the facial nerve. The vestibular aqueducts are not enlarged. The vestibule and semicircular canals are well seen and appear normal.
- Initially diagnosed with a moderate to severe mixed hearing loss which was progressive

10 months post-implant

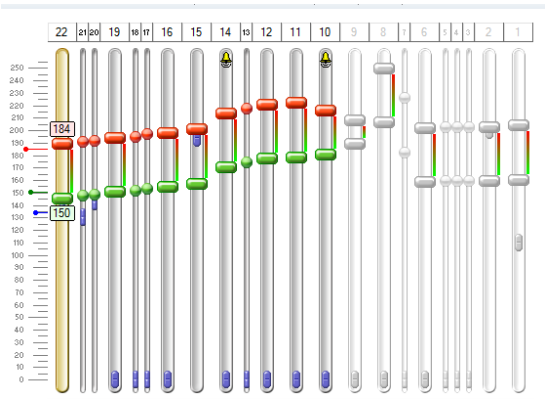


AidedTesting	Right CI
NU-CHIPS	92%

One month later…

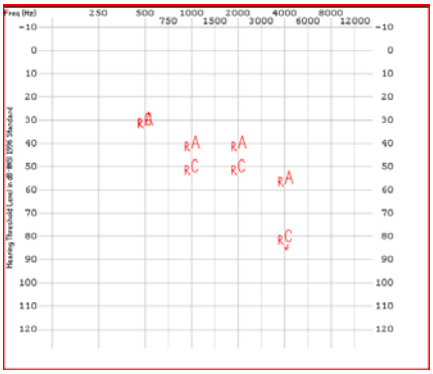


Case 8: Stapes Gusher



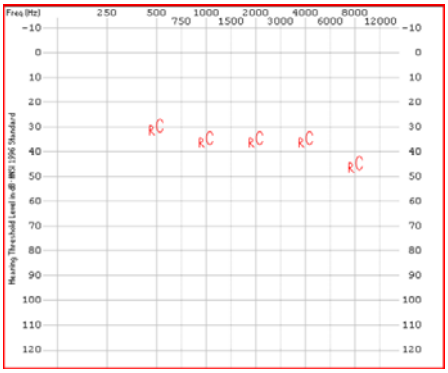
C	184	189	193	196	209	216	217	211	203	245	197	198	199
T	150	155	159	162	175	182	183	185	194	211	163	164	165
MRK	134	0	0	194	0	0	0	0	0	0	0	200	110
AE	22	19	16	15	14	12	11	10	9	8	6	2	1
Gain	0	0	0	0	0	0	0	0	0	0	0	0	0
PW	25	25	25	25	25	25	25	25	25	25	25	25	25
DR	34	34	34	34	34	34	34	26	9	34	34	34	34
UF	313	1063	2063	2563	3188	5063	6313	7938	0	0	0	0	0
LF	188	813	1688	2063	2563	4063	5063	6313	0	0	0	0	0

After Programming



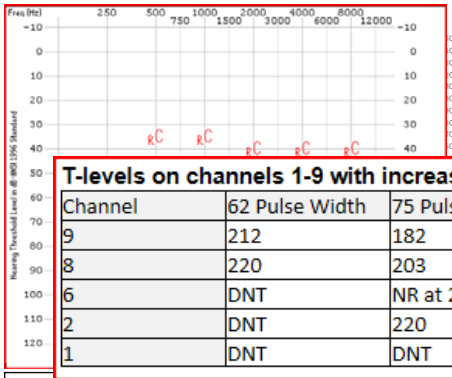
Aided Testing	Right CI
NU-CHIPS (Old Map)	92%

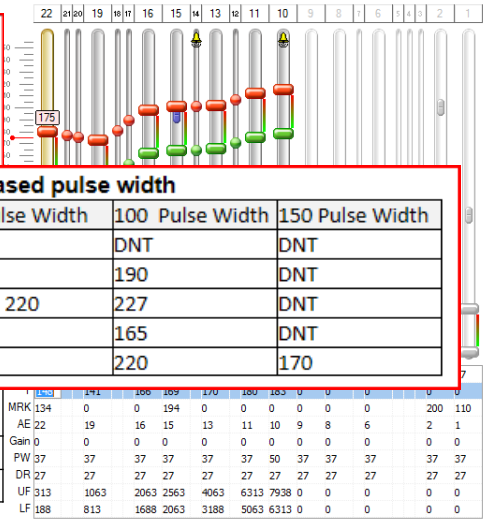
One month later…



Aided Testing	Right CI
NU-CHIPS	92%

Case 8: Stapes Gusher





T-levels on channels 1-9 with increased pulse width

Channel	62 Pulse Width	75 Pulse Width	100 Pulse Width	150 Pulse Width
9	212	182	DNT	DNT
8	220	203	190	DNT
6	DNT	NR at 220	227	DNT
2	DNT	220	165	DNT
1	DNT	DNT	220	170

Aided Testing	Right CI
NU-CHIPS	86%
MLNT Easy (Recorded)	75%

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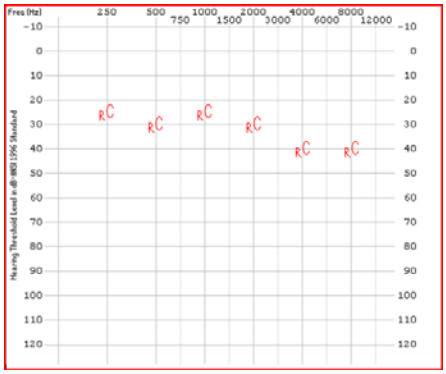
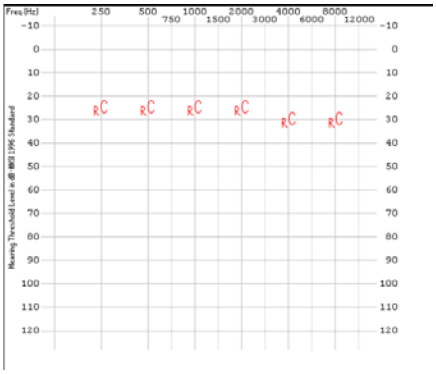
One Month Later…



Aided Testing	Right CI
MLNT Easy (Recorded)	93%

One Year Later…



Aided Testing	Right CI	Left HA	Bimodal
SRT	25	DNT	DNT
MLNT Hard (Recorded)	91%	DNT	67% (attention)

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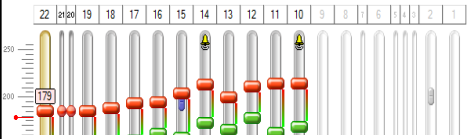
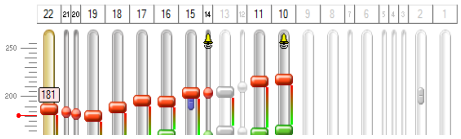
Case 8: Stapes Gusher





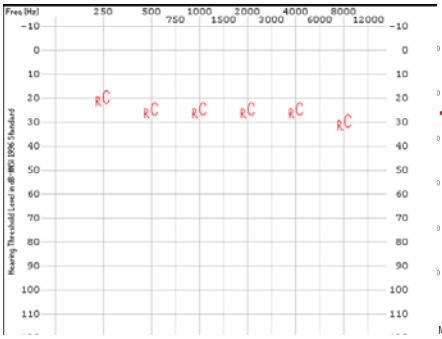
T-Levels on channels 12 and 13 with increased pulse widths

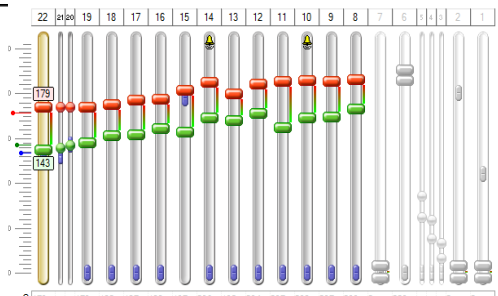
Channel	50 Pulse Width	62 Pulse Width	75 Pulse Width	88 Pulse Width
12	192	178	178	171
13	No Response	Not recorded	187	184

C	179	179	182	187	188	197	206	193	204	207	208	2	2	2	2	2
T	143	151	159	160	167	163	179	171	184	168	174	0	0	0	0	0
MRK	134	0	0	0	0	194	0	0	0	0	0	0	0	200	110	
AE	22	19	18	17	16	15	14	13	12	11	10	9	8	6	2	1
Gain	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
PW	37	37	37	37	37	37	37	88	88	37	37	37	37	37	37	37
DR	36	28	23	27	21	34	27	22	20	39	34	2	2	2	2	2
UF	313	1063	1313	1688	2063	2563	3188	4063	5063	6313	7938	0	0	0	0	0
LF	188	813	1063	1313	1688	2063	2563	3188	4063	5063	6313	0	0	0	0	0

C	181	174	183	190	189	197	198	209	211	23	23	23	23	23	
T	133	146	147	154	166	160	170	168	171	0	0	0	0	0	
MRK	134	0	0	0	0	194	0	0	0	0	0	0	200	110	
AE	22	19	18	17	16	15	13	11	10	9	8	6	2	1	
Gain	0	0	0	0	0	0	0	0	0	0	0	0	0	0	
PW	37	37	37	37	37	37	37	37	50	37	37	37	37	37	
DR	43	28	36	36	23	37	28	41	40	23	23	23	23	23	
UF	313	1063	1438	1938	2563	3438	0	6063	7938	0	0	0	0	0	
LF	188	813	1063	1438	1938	2563	0	4563	6063	0	0	0	0	0	

Case 8: Stapes Gusher

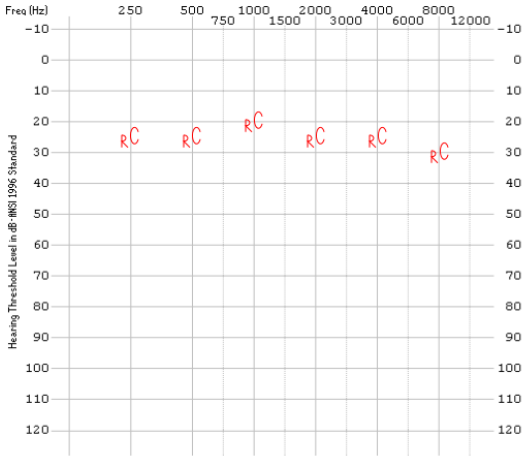




Aided Testing	Right CI	Left HA	Bimodal
SRT	20	DNT	DNT
MLNT Hard	91%	DNT	83%
LNT Hard	60%	DNT	72%
PBK	52%*	DNT	48%*

C	179	179	182	187	188	197	206	193	204	207	208	207	209	2	220	2	2
T	143	151	159	160	167	163	179	176	184	168	179	180	189	0	220	0	0
MRK	134	0	0	0	0	194	0	0	0	0	0	0	0	0	0	200	110
AE	22	19	18	17	16	15	14	13	12	11	10	9	8	7	6	2	1
Gain	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
PW	37	37	37	37	37	37	37	88	88	37	37	100	100	150	100	37	37
DR	36	28	23	27	21	34	27	17	20	39	29	27	20	2	0	2	2
UF	313	813	1063	1313	1563	1938	2313	2813	3438	4188	5188	6313	7938	0	0	0	0
LF	188	563	813	1063	1313	1563	1938	2313	2813	3438	4188	5188	6313	0	0	0	0

Case 8: Stapes Gusher (Current Performance)



Aided Testing	**Right CI**	**Left HA**	**Bimodal**
SRT	20	DNT	DNT
MLNT Hard	83%	DNT	91%
LNT Hard	72%	DNT	76%
PBK (recorded)	56%*	DNT	68%*
PBK (MLV)	68%	DNT	76%

Pediatric Hybrid

- Disclaimer: Hybrid cochlear implants are <u>not</u> approved for use in the pediatric population
- However, "hybrid" can refer to:
  - The actual implant (short array)
  - The fitting approach (traditional implant array)
- Patients (ped and adult) are being implanted with standard arrays, and coming out of surgery with significant residual hearing
- What do we do with these?

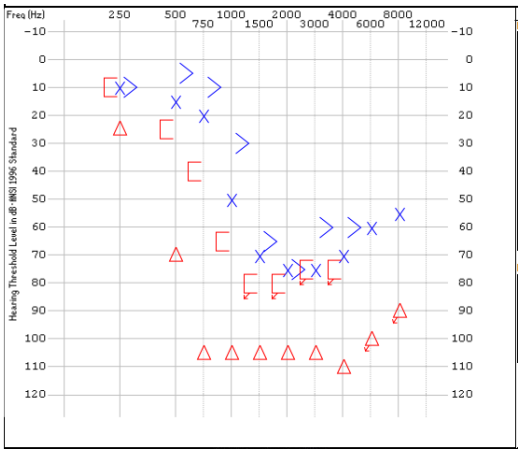
Case 9: Simulated Hybrid

- Etiology unknown
- Identified at 3 years of age and fit at 5 years of age
- Progressive sensorineural hearing loss (right>left)
- Fit with Bi-CROS at 10 years

Case 9: Simulated Hybrid (12 years of age)

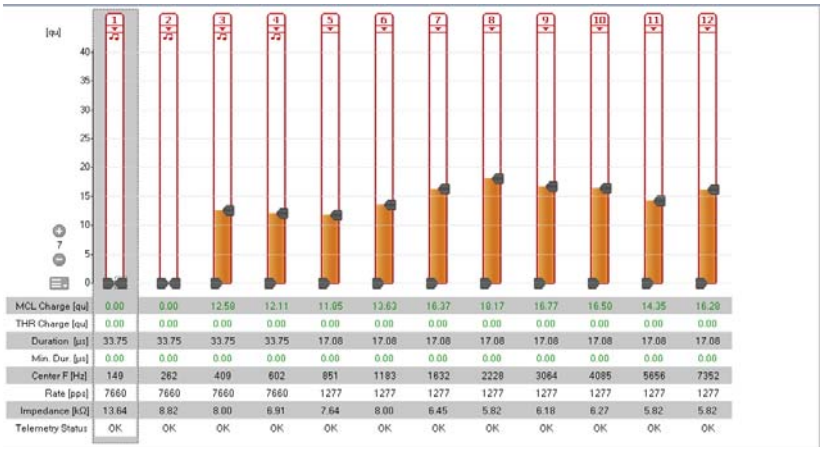
Pre-Op Aided Speech Perception Testing	Aided: Binaural	Aided: Bi-CROS	Aided: Left	Aided: Right (Loaner Hearing Aid)
W22 (MLV)	DNT	DNT	88%	32%
W22 (Recorded)	DNT	DNT	65%	40%
CNC (Recorded)	DNT	DNT	52%	28%
BKB SIN (Recorded)	4.5 dB SNR	3.5 dB SNR	2.5 dB SNR	9.5 dB SNR

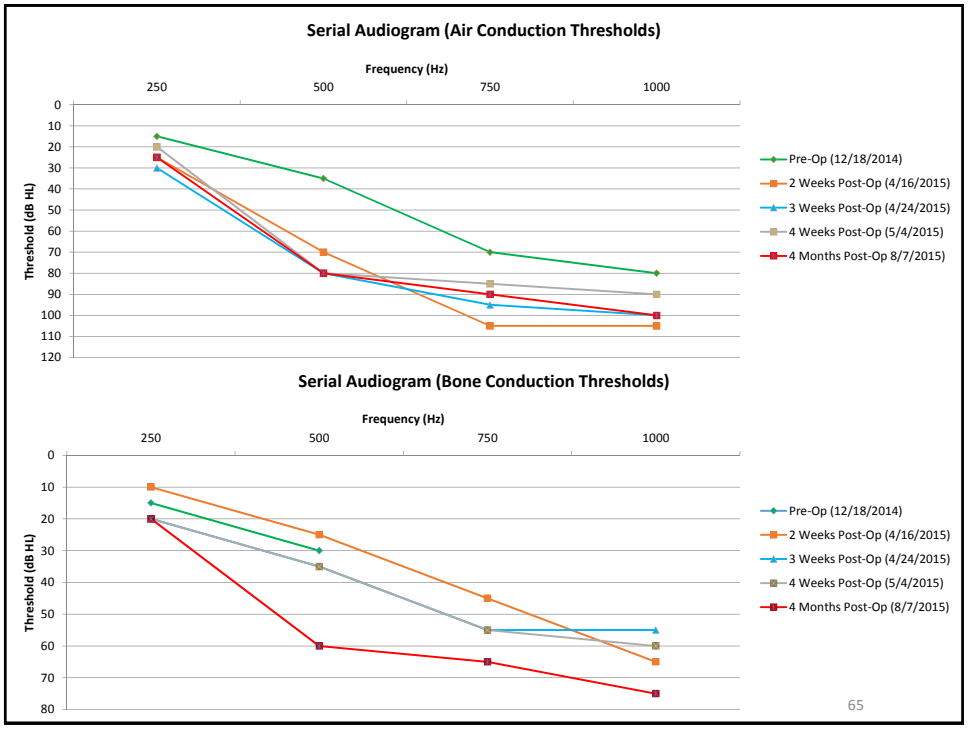
Case 9: Simulated Hybrid Post-op Unaided Testing



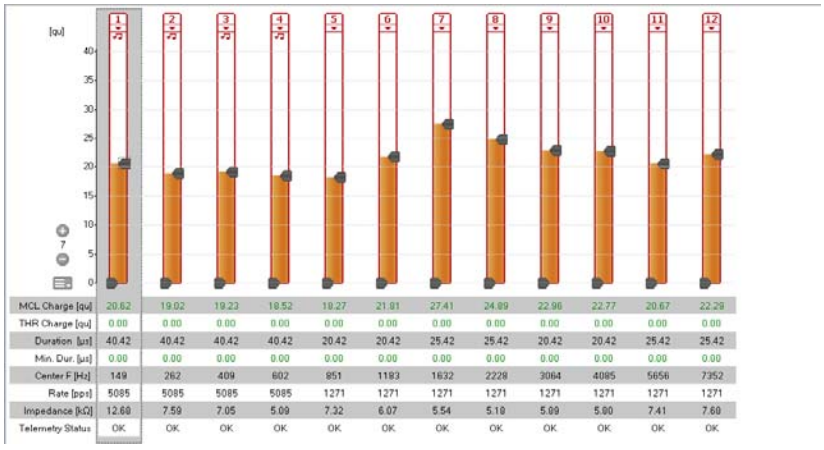
Post-Op Unaided Speech Perception Testing	Right (CI)	Left
SRT	60 dB HL	25 dB HL
CNC (Recorded)	0%	64%

Case 9: Simulated Hybrid Initial Activation

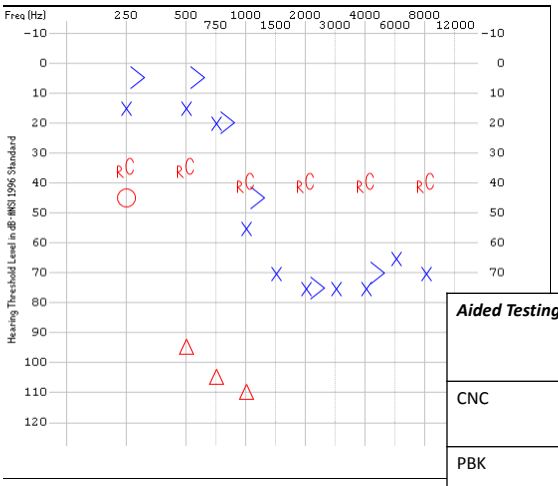




Case 9: Simulated Hybrid
4 Months Post



6 Months Post



Unaided	Left Ear	Right Ear
SRT	30	DNT
CNC	71%	DNT

Aided Testing	Right Ear Cochlear Implant	Left Ear Hearing Aid	Bimodal
CNC	DNT	80%	DNT
PBK	Recorded: 16% MLV: 84%	DNT	DNT
BKB-SIN	DNT	DNT	+ 3.5 dB SNR

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Case 10: Tinnitus and CI

- Etiology Kabuki Syndrome
- Identified at 5 years with a mild sensorineural hearing loss which was progressive (right>left). Fit with bilateral hearing aids at that time
- Used bilateral hearing aids for 10 years, then discontinued right aid due to lack of benefit.
- Two years later fit with right hearing aid to attempt to mask tinnitus

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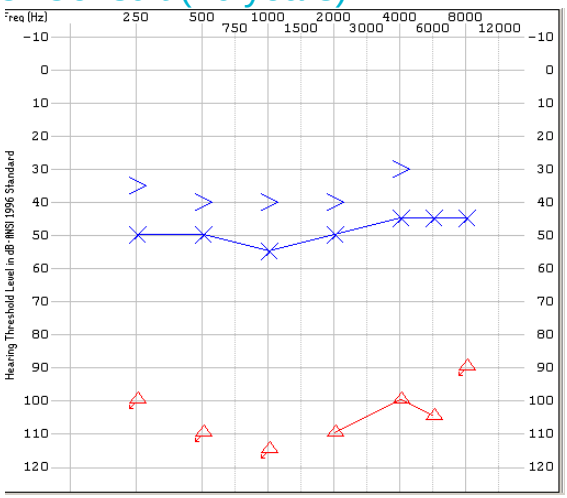
Cincinnati Children's
changing the outcome together

Case 10: Tinnitus and CI

CI Consult (16 years)



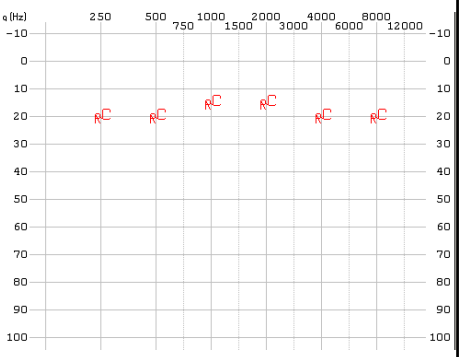
Pre-Op Unaided Speech Perception	Left Aided
CID W22 at 50 dB HL	100%
CID W22 at 35 dB HL	48%

Tinnitus Questionnaire		
Area of Concern	**Patient Score**	**Total Possible**
Tinnitus associated with use of hearing aids	17	20
Annoyance Value	9	20
Frequency	8	15
Total	34	55

Post Implant Tinnitus

Tinnitus Questionnaire			
Area of Concern	Pre-Implant	Post-Implant	Total Possible
Tinnitus associated with use of hearing aids	17	4	20
Annoyance Value	9	11	20
Frequency	8	9	15
Total	34	24	55

Current Performance



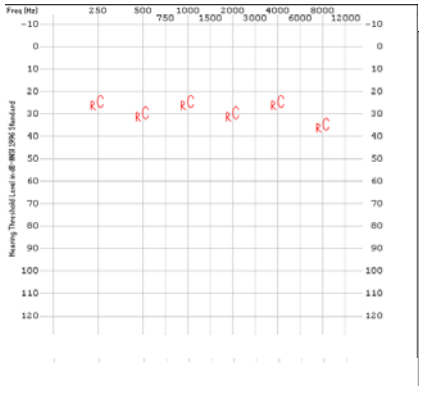
Aided Testing	CI Only	Bimodal
CNC Words	92%	DNT
CNC Phonemes	94%	DNT
BKB-SIN in noise	+4 dB SNR	-1 dB SNR

Case Study 11: remote access

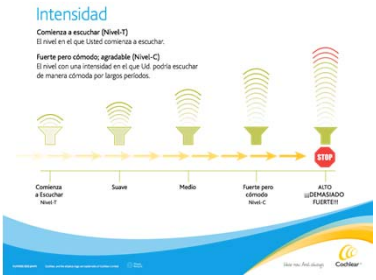
- 23 year old male
- Long-standing severe to profound SNHL
- Uses hearing aids intermittently due to days where sound quality is very poor
- MRI and CT scans: “findings compatible with bilateral cochlear nerve deficiency”
- Interested in CI, with appropriate expectations
- Home: Dominican Republic

Case Study 11: remote access

- Right cochlear implant
- Had initial activation and first three follow up appointments in Cincinnati
- Trained on what to expect for follow-up at home
- Provided home therapy excercises



Case Study 11: Remote Access



"Success" defined

a : degree or measure of succeeding

b : favorable or desired outcome;

also **:** the attainment of wealth, favor, or eminence

- Merriam-Webster Online

How do you want to define "success"?
How do the parents want to define "success"?
Do they match up?

Issues to Consider

- In more complex cases, hearing loss may not be the first priority
- Some disabilities are not easily identifiable at the time of consideration for candidacy
- Even in children without additional disabilities, outcomes depend on significant factors such as chosen mode of communication

> "Despite the best efforts of many professionals, it is often difficult to diagnose learning disabilities, reduced cognitive function, and soft neurologic deficits in very young children…"
>
> Walzman, 2000

Beyond audiograms and speech perception measures…

Some thoughts and questions to ponder:

- Is there additional information that should be considered for closer analysis?
- Are we already getting the information without evaluating it's value?
- Due to age, attention, location, etc., many patients are not able to provide accurate feedback while the audiologist programs their cochlear implant, so we have to consider several other **tools**…

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